394 How a team adherence challenge led to increased communication between teenage patients and the CF team

P. Cole[1], F. Lindsay[1], P. Singleton[1], J. Pagan[1], B. Ashby[1]. [1]*Teesside CF service, Middlesbrough, United Kingdom*

Background: The CF team undertook an adherence challenge to experience aspects of CF management. Ensuing discussions with teenagers made it apparent that many had experiences they wanted to share relating to developing coping mechanisms and accepting CF into their lives. They also wanted to see other teenagers with CF and understand how they coped. Segregation for infection control has deprived them of direct contact. They suggested a DVD illustrating some young people's perspectives would be a useful tool.
Method: Teenagers and young people were invited to participate in the planning and production of a DVD. Most were enthusiastic and wanted to be involved even if they didn't want to be filmed. Teleconferencing facilities allowed safe communication between the patients. The local university provided technical expertise and it was financed from donations.
Outcome: A 14 minute DVD was produced featuring 4 patients and 3 team members. Teenagers demonstrated how they balanced daily activities with the time needed for their CF care. They felt this achieved their aim of presenting a positive, non-clinical image without ignoring the challenges of coping with CF. Team members reflected on the challenge and how it altered their approach to teenagers with CF.
Teleconferencing proved a successful method of empowering the patients to express their feelings and ideas about living with CF.
Reflections: The process has encouraged discussions and given the team the opportunity to probe issues that are difficult to address in clinic. Most young people are yet to see the DVD to give feedback. It will be shown at the parents group to give them some ideas about their children as teenagers. Future plans to improve communication include more teleconferencing and a DVD featuring younger children.

395 Newborn screening in the northern region, difficulties and dilemmas

A. Sewell[1], C. Sharpe[1], J. Bloomer[1], D. Spencer[1]. [1]*Paediatric Cystic Fibrosis Unit, Royal Victoria Infirmary, Newcastle Upon Tyne, United Kingdom*

In November 2006 newborn screening for CF was rolled out across the northern region of the UK. This programme follows the pathway developed by the UK Newborn Screening Programme.
We report the difficulties and dilemmas that have arisen so far.
To date 8 babies have been diagnosed with CF. 16 carriers have been identified, of which 3 initially had positive screening tests. These 3 cases, who had one identified mutation and a second raised IRT, highlighted the difficulties in sweat testing small babies in order to confirm their diagnosis.
Offering sweat tests to siblings following a positive CF newborn result has lead to 'well' siblings being diagnosed. Families taking up cascade genetic screening has identified extended family members as having CF. These issues have added further to the stress felt by families and contributed to difficulties in coming to terms with the diagnosis.
The death of a baby with Downs syndrome diagnosed with CF on newborn screening, proved difficult for all involved and raised many ethical dilemmas.
The national programme identifies Health Visitors as being the appropriate professional to give results to families. However, in this region it has become apparent that some may not have the skills and knowledge with which to undertake this role. This has led to some families being misinformed about the screening result and its implications. In order to address this, multi-disciplinary training sessions have been established to assist the Health Visitors in delivering reults.
The benefits of early diagnosis and prompt treatment are well documented. Newborn screening in our region is still in the early stages but has already presented many challenges which will hopefully help to prepare us for the further dilemmas ahead.

396 Influence of segregation policy on daily functioning of adult cystic fibrosis patients

E.A. Boerhof[1], W. Doeleman[2], G. Kempen[3]. [1]*Pulmonology, University Medical Center, Utrecht, Netherlands;* [2]*Rehabilitation and Sports Medicine, University Medical Center, Utrecht, Netherlands;* [3]*School for Public Health and Primary Care, University Maastricht, Maastricht, Netherlands*

Introduction: Literature shows that segregation of CF patients reduces the chance for cross-infection but may affect quality of life and negatively influence daily functioning. At the UMCU, segregation policy for adult CF patients was implemented from 15 October 2005. This study investigated changes in social contacts between CF patients before and after the introduction of the segregation policy and how this affected their health-related quality of life (HRQoL).
Methods: Two sub-studies were carried out. The first comprised statistical analyses of CF questionnaires (CFQ) patients completed before and after implementation of the segregation policy. We included 162 questionnaires of 81 patients for analyses. Semi-structured interviews were carried out with 10 patients to investigate their experiences with the segregation policy.
Results: This study showed a significant decrease in three out of twelve HRQoL domains after implementation of the segregation policy. These domains are vitality, treatment burden and emotional state. Median of FEV1 (% of expected) was 58 (range 27–113), median of age was 25 (range 18–52). The segregation policy has negative impact when patients are admitted to the hospital. Most patients feel locked up in a room and they experience wearing a mouthmask as very stigmatizing and unpleasant. Median of FEV1 of interviewed patients was 35 (range17–95) and median of age 26.5 (range 20–49).
Conclusion: Segregation policy threatens HRQoL and contacts with fellow patients, but it is the patients personal decision to meet fellow patients outside the hospital.

397 Evaluation of "patient readiness" to graduate from pediatric to adult health care

A.M. Gravelle[1], A.G. Davidson[1]. [1]*Cystic Fibrosis Clinic, B.C. Children's Hospital, Vancouver, BC, Canada*

"Graduation" from pediatric to adult clinics is an important milestone for Cystic Fibrosis (CF) patients. Adolescent transition protocols have been developed to help prepare youth for the adult setting, but these are relatively new and there is little data to evaluate their efficacy. We have recently created and tested a "readiness to graduate" questionnaire in order to evaluate our own comprehensive adolescent transition program.
The questionnaire is CF-specific based on a generic chronic illness adolescent transition framework[1]. Answering "yes" to a question indicates readiness; "somewhat" indicates more education/intervention is needed; and "no" indicates a patient is not prepared for adult healthcare in that particular area.
The questionnaire was administered by our CF Nurse Clinician to 12 CF patients, ages $17\frac{1}{2}$ to 18 years, who had participated in our transition program since age 11–12 yrs, and were about to "graduate" to the adult CF clinic. Eight youth demonstrated a strong sense of readiness to transfer to an adult CF clinic; 3/12 had an acceptable level of readiness; and one (who had frequently missed clinic appointments) was "not ready". The most commonly failed questions dealt with filling a prescription; medical insurance; and proper name and dosage of all medications.
As a clinic with a longstanding comittment to adolescent transition care, these results provide assurance that our CF adolescent transition program is largely meeting its goals. They also identify the need for extra attention in certain areas. The tool was quick and easy to use, and very useful to youth and their parents as a concrete indicator of readiness to "graduate".

Reference(s)

[1] Paone M, et al. On TRAC: a transition service at B.C. Children's Hospital, Vancouver, 1997.